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INHERITED SYPHILIS AND BLUE SCLEROTICS.

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With Dr J. D. Rolleston's compliments

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INHERITED SYPHILIS AND BLUE SCLEROTICS.*

By J. D. Rolleston, M.D.,
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A male infant, aged 5 months, an only child, was admitted to the Grove Hospital for nasal diphtheria on November the 15th, 1910. The father had contracted syphilis about six years previously, and had undergone only a few months' treatment. Six months after marriage and one year before the baby's birth the mother developed "a poisoned lip," probably a hard chancre, of which the scar is still visible in the centre of the lower lip. Its real nature does not seem to have been recognised, as she received only local treatment on seeking advice at a hospital.

When three weeks old the baby lost all power in his left arm, but recovered the use of the limb in the course of a month without specific treatment, and when six weeks old had "thrush at both ends," which was probably mucous tubercles in the mouth and round the anus.

Condition on admission.—Atrophic, fair-haired infant, weight 8 lb. 7 oz., constantly crying. Marked prominence of scalp veins, bulging forehead, umbilical and right inguinal hernia. Crusts in right nostril from which diphtheria bacilli were cultivated. No eruption. Spleen and liver not enlarged. Heart normal.

Both sclerotics show a uniform pale blue coloration; irides grey, embryontoxon at the margin of each cornea.

During the child's stay in hospital the blue coloration of the sclerotics was more obvious on some days than on others.

The mother, aged 27 years, a woman of fair complexion, presented an almost identical condition of the sclerotics, but the coloration was somewhat deeper, more nearly approaching a leaden hue.

An arcus senilis was present in each eye. Beyond occasional smarting on exposure to wind and lacrymation following prolonged accommodation, her eyes had not caused her any trouble. She stated that her sister, a fair-haired girl, aged 16 years, presented the same condition, as did also their grandmother, now dead. Their two brothers, aged 37 and 22 years respectively, were not affected.

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* Case shown at the Royal Society of Medicine (Section for the Study of Disease in Children) on March the 24th, 1911.

The child's nasal diphtheria cleared up under 4000 units of antitoxin, but diphtheria bacilli persisted in the nose until February, 1911. Four days after admission periostitis of the upper end of the left ulna developed, but subsided in a few days after administration of hydrarg. c. creta gr. ½ *bis die*. Beyond some erosions round the anus and paronychia of the right ring finger nothing further of note occurred until January the 12th, 1911, when the left arm was found to present the flail-like attitude characteristic of Parrot's syphilitic pseudo-paralysis. On palpation the arm was found to be excessively tender, and definite crepitus was felt at the junction of the upper and middle third of the humerus. The temperature was 101° F. for two days, and then became normal.

The limb was put up for a fortnight in a cardboard splint, and the mercury, which two days before the fracture had been reduced to gr. ¼ *bis die* owing to loose stools, was increased on January the 21st to gr. ½ *b.d.* and on February the 4th to gr. ½ *ter die*. On January the 28th when the splint was removed there was good union, and well-marked callus was felt. Active and passive movements were free. Subsequent recovery was uneventful, and the child was discharged in good health on March the 9th, 1911.

The principal features of interest in the case are the extra-genital infection of the mother, the spontaneous fracture of the humerus in the child, and the condition of blue sclerotics in three generations. Although no Wassermann's reaction was performed, the history of the case and the position of the scar made it practically certain that the lesion on the lip had been a chancre.

Spontaneous fractures, or, to use Broca's more accurate term, "pathological fractures," are comparatively uncommon in syphilis, especially fractures of the shaft as distinct from separation of the epiphyses, and still more exceptional is it for the fracture to be limited to a single bone instead of the lesions being multiple, as in most of the recorded cases. This was probably due to the adoption of specific treatment, as shortly after admission, before mercury had been given, the child developed periostitis of the ulna, thus indicating that the osseous system showed a special tendency to be involved.

The symptom of persistent crying, to which attention has recently been drawn by Comby and Sisto as a phenomenon of inherited syphilis, was probably due to pains in the bones.

The occurrence of the fracture during mercurial treatment was probably due to the doses being too small, as rapid recovery ensued on increase of the dose.

The case is illustrative of the favourable course of pathological fractures in inherited syphilis, provided suitable treatment is adopted. All authorities now hold this view, but Parrot, who first described syphilitic pseudo-paralysis, regarded it as a very unfavourable sign as all his cases died.

If the history could be trusted the same arm had already been affected when the child was three weeks old, and recovery had taken place without specific treatment. A similar case of spontaneous recovery from syphilitic pseudo-paralysis has been recorded by Cadet de Gassicourt, and Gonez has collected five other cases in which improvement occurred before mercury was administered.

It should be noted that the humerus is the most frequent site of pathological fractures in syphilis, as it was affected in twenty-two out of sixty-four such cases collected by Frangenheim.

The condition of blue sclerotics as a congenital disease was first described by von Ammon in 1841 in the following terms : " Congenital diseases of the sclerotic are rare. . . . Of importance is a peculiar whitish-blue coloration of this membrane occasionally met with, when the whole development of the eye is retarded. The sclerotic in such cases appears thin and almost transparent. I have seen it also in congenital hydrophthalmos. . . . Similar thinness occurs in patients suffering from congenital heart disease. In that case the sclerotic is dark blue, this being due partly to the thin condition of the membrane and partly to an accumulation of venous blood and a large mass of pigment in the eye."

Von Ammon's description is accompanied by two illustrations (Tab. xv, figs. 2 and 3), the first representing the blue coloration in a case of congenital hydrophthalmos, and the second in a case of congenital morbus cordis.

After von Ammon little notice seems to have been taken of the anomaly until 1903, when Dr. Leslie Buchanan, of Glasgow, described a case in a girl, aged 9 years, whose left eye he examined after excision for an injury. He found that the cornea and sclerotic were unusually thin, the cornea being three fifths and the sclerotic one third of its normal thickness. Histological examination showed that the fibres of the cornea and sclerotic were of about normal size but unusually few in number. The anterior elastic lamina was entirely absent.

In a paper entitled " A Congenital Anomaly of the Sclera : Pseudocoloboma," to which Mr. Sydney Stephenson has kindly drawn my attention, Dr. Percival J. Hay, of Sheffield, in 1907, described a condition resembling that under discussion, but differing from it in that the thinning of the sclerotics, instead of being uniform and symmetrical, was in the right eye represented by a triangular area on either side of the cornea, and on the left confined to an area on the temporal side only. The case occurred in a still-born child, the subject of many other congenital deformities. There was no family history. It was not till 1908 that the hereditary transmission of blue sclerotics was first mentioned. In that year A. Peters, of Rostock, recorded cases in four generations. Four of his patients showed a typical embryontoxon. Peters regarded the condition as due to an abnormally thin or abnormally transparent sclerotic. In 1910 Mr. Sydney Stephenson described the condition affecting twenty-one out of thirty-two members belonging to four generations of one family. In his cases, as in mine, the inheritance was through the females, and the complexion in general was fair. In two cases the presence of an arcus senilis or an embryontoxon was noted. Subsequent investigation enabled Mr. Bishop Harman to add another generation to this family, so that a total of fifty-five members was reached, of whom thirty-one showed the same congenital peculiarity with individual differences.

It did not occur to me that there was any connection between the pathological fracture of the child's humerus and its blue sclerotics until, while looking up the literature on abnormalities of the sclerotic in the Surgeon-General's Index Catalogue I came across a paper by Dr. Eddowes, entitled "Dark Sclerotics and Fragilitas Ossium," where he described some cases running in families in which the two conditions were associated. Dr. Eddowes suggested that the transparency of the sclerotics indicated a want of quantity or quality in the fibrous tissue forming the framework of the various organs of the body, which therefore accounted for the want of spring and toughness in the bones. Dr. Eddowes has kindly informed me that he never even suspected inherited syphilis in his cases.

Possibly the deficiency of fibrous tissue in the present case as manifested by the blue coloration of the sclerotics may have been a contributing factor together with syphilis in the production of the fracture. There was, however, no history of fragilitas ossium in any other member of the family.

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